Repair of the Regurgitant Bicuspid or Tricuspid Aortic Valve
Background, Principles, and Outcomes

Fabian A. Kari, MD; Matthias Siepe, MD; Hans-Hinrich Sievers, MD; Friedhelm Beyersdorf, MD

The treatment of severe aortic valve regurgitation (AR) has been moving ahead quickly and has undergone a fundamental transition over the past few decades. Mechanical or bioprosthetic aortic valve replacement has been the standard of care for many years for most types of AR. Although still restricted to patients with well-defined pathologic aortic valve and root anatomy, the number of accepted indications for reconstructive aortic valve surgery is growing.

The cusp tissue, aortic annulus, tissue of the sinus of Valsalva, commissures, subcommissural tissue, and sinotubular junction constitute the functional unit of what is generally called the aortic root. Reconstructive surgery for severe aortic valve insufficiency today includes a wide variety of procedures on all of those components, from simple cusp plication to complex valve-sparing aortic root replacement (V-SARR). It is ultimately aimed not only at improving symptoms of congestive heart failure and increasing life expectancy but also at giving the patient a chance to live a normal life because it renders lifelong anticoagulation unnecessary. V-SARR procedures have recently been discussed not only as adequate correction for aneurysms but even as supplementary procedures to prolong the longevity of a valve repair.

Our article gives an overview of the background and principles of reconstructive aortic valve procedures for AR in the setting of both a bicuspid and a tricuspid aortic valve. It explains the principles, describes the techniques, and reviews the reported current outcomes after reconstruction of the different components of the aortic root. This overview should help clinicians evaluate the status and relevance of frequently discussed current techniques for the treatment of patients with different types of thoracic aortic aneurysms and AR.

Advantages of Aortic Valve-Sparing Surgery: Why Repair?
The need for lifelong anticoagulation remains the main drawback of prosthetic aortic valve replacement for many young patients living an active lifestyle. In addition to sparing the patient’s native valve and thus avoiding anticoagulation, a valve-sparing reconstructive procedure has several theoretical advantages. First, autologous valve cusp tissue may be linked to less structural deterioration over time than is the case with bioprosthetic pericardial valves. The younger the patient is and the better the left ventricular ejection fraction is, the earlier an aortic bioprosthesis degenerates and mandates reoperation. Focusing on AR, one must, however, differentiate among type 1 AR caused by isolated annular dilatation, type 2 AR resulting from cusp prolapse, and combinations of these types. Cusps of an aortic valve with pure type 1 AR may be completely normal on the cellular and molecular levels. In contrast, prolapsing cusps in type 2 AR may be affected by inherent structural and physiological derangements on the cellular level. Worse midterm and long-term results of reconstructive approaches can thus be expected with these valves. Hemodynamics of the left ventricular outflow tract, aortic valve, aortic root, and ascending aorta probably change significantly after implantation of a bioprosthesis. Although it has yet to be proven, aortic valve reconstruction or V-SARR may preserve the patient’s individual ventricular and aortic hemodynamic profile.

Reconstructive aortic valve procedures replace aneurysmal tissue and correct for AR. These procedures have long consisted of a highly variable set of markedly different techniques. Included are procedures on the free margins of the cusps and the cusps themselves, as well as procedures on other aortic root components like the commissures, the subcommissural (sometimes called interleaflet) triangles, and aortic sinus tissue (reimplantation, remodeling). For every component of the functional unit of the aortic root—specifically the annulus, cusps, commissures, sinuses, and the sinotubular junction—reconstructive approaches have been described and performed, and most of them aim at correction of AR.

The decision to perform a valve repair, the choice of technique, and the combination of different techniques, as is often necessary, are all determined by the individual surgeon. These choices are based largely on personal experience, subjective judgment, and individual preference. Nevertheless, although still far from being uniform among countries or even within the same institution, repair of the regurgitant bicuspid (BAV) or tricuspid (TAV) aortic valve has slowly evolved in recent years toward a more consistently characterized and more standardized armamentarium of strategies.

Several attempts to systematically characterize the mechanisms of AR in TAV or BAV that focus on relevance for surgical repair rather than on morphology have contributed to this evolution. Lesions of the cusps and other components of

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the aortic root that contribute to AR and need to be surgically addressed singly or in combination are now better understood and described.7 For BAVs, a recently introduced set of categories and subcategories facilitates unambiguous description and communication of pathological cusp configurations.8 All of these contributions have led to a better understanding of the aortic valve characteristics that, apart from the patient’s clinical characteristics, make V-SARR (Figure 1) an appropriate therapeutic option. The emphasis on mechanisms of AR helps to determine the utility of additional repair of the cusps and use of other adjunctive modifications. This function-oriented approach also leads to a better understanding of which technique is most suitable for a particular mechanism of AR.

**Functional Unit of the Left Ventricular Outflow Tract and Supravalvular Aorta**

The aortic and mitral valves are positioned within the left ventricular orifice in close proximity and anatomic continuity. The subvalvular membrane attaches both valves to the rim of the left ventricular orifice, with the subaortic curtain being supported by both the right and left fibrous trigones. Reconstruction of the aortic valve is possible without interfering with mitral anatomy, however, because the right and left fibrous trigones and the subaortic curtain preserve a minimal but sufficient distance between the valves. Thus, even plicating the entire aortic annulus usually does not interfere with mitral anatomy.

The left ventricular outflow tract, aortic valve, and aortic root constitute a finely tuned functional complex, the details of which are not yet completely understood. Blood flow within the left ventricular outflow tract is rotational; this blood flow pattern persists throughout the ascending aorta and extends into the aortic arch. Physiological flow patterns constituting a vortex within the sinuses of Valsalva may be important for the integrity of the aortic wall and normal cusp opening and closing movements. After replacement of the aortic valve, this finely tuned hemodynamic unit is likely to undergo substantial changes, starting at the level of the implanted tilting-disk mechanical or trileaflet porcine or ovine bioprosthesis. Many assume that a V-SARR procedure combined with cusp repair should help to preserve a close-to-normal hemodynamic situation because the anatomy and the movement of the ventriculo-aortic interface remain physiological.

**Mechanisms of AR**

El Khoury and colleagues2,7,9 have introduced a classification of AR that characterizes regurgitation mechanisms with the hope of clarifying the optimal surgical treatment strategy. Type I AR is defined as regurgitation in the presence of normal cusp mobility. Dilatation of components of what is called the functional aortic annulus—the subannular aortoventricular junction, the mid aortic root, or the sinotubular junction—is the causative mechanism for type I AR. In contrast, cusp mobility is abnormal in types II and III; it is severely enhanced in type II and severely restricted in type III. In type II AR, cusp tissue redundancy is evident, resulting in prolapse of 1 or more cusps into the left ventricular outflow tract, below a level mandatory for physiological coaptation. In type III AR, retraction of tissue as a result of fibrosis or calcification results in restricted cusp mobility. A combination of different mechanisms may lead to severe AR and may require the application of a combination of repair strategies targeting different components of the aortic root, for example, in the case of a BAV that has a dilated or severely elliptical

![Figure 1](image-url)

Figure 1. Modifications of the Tirone David valve-sparing aortic root replacement procedure. Types II, II, IV, and V represent refinements and additions to the original type I procedure using a single relatively small graft. These modifications aim at more physiological sinus of Valsalva function by creating a new sinotubular junction and by downsizing the annulus to a higher degree compared with the sinus level. Smod indicates Stanford modification.
Principles of Aortic Valve Repair
With or Without V-SARR

Sufficient coaptation of the cusps needs to be established without sacrificing too much cusp mobility; an immobile cusp can lead to AR or aortic stenosis either immediately or late postoperatively. The goal of aortic valve repair is to achieve a homogeneous coaptation level for all cusps, ideally above the aortic annulus, similar to the approach established for repair of the mitral valve. The optimal coaptation level for the aortic valve is thought to be at about the midpoint of the height of the aortic sinuses. If a nonprolapsing cusp is present, its coaptation level can be used as the ideal. If not, the midsinus point can be used as reference, or with the use of quantification tools like the Schaefer caliper, a coaptation height of 8 to 10 mm can be achieved. The effective cusp height is defined as the vertical distance from the lowest point of the cusp free margin to the level of the aortic annulus, and it should be >8 mm after repair.

Especially in BAVs, the mechanism of AR may be multifactorial, not purely a problem of cusp mobility or the quantity of cusp tissue. The importance of addressing both components of AR. Cusp pathology should be dealt with using techniques like central or peripheral free-margin plication, raphe shaving, triangular raphe resection, or free-margin resuspension, correcting for cusp tissue redundancy, excessive mobility, or impaired mobility. Figure 2 gives an overview. The functional aortic annulus, consisting of the subannular aortoventricular junction and the sinotubular junction, also needs to be addressed. Aortic root dilatation may contribute to the severity of AR. Combinations of subannular plication techniques and techniques replacing the sinus tissue for patients requiring annular downsizing in the presence of a root aneurysm are performed by several groups.

Adding a V-SARR even if sinuses are normal has been proposed as a measure to increase long-term durability of aortic valve cusp repair. This is a novel and controversial extension of the spectrum of indications for a procedure originally designed for replacing aneurysmal root and ascending aortic tissue. The long-term benefit of an additional procedure as complex and challenging as a V-SARR is debatable, especially considering the fact that early mortality and midterm and long-term functional outcomes differ substantially between centers. An additional V-SARR performed in a low-volume center may expose the patient to a significantly increased and potentially unnecessary risk.

Bicuspid Aortic Valve

BAV is the most common congenital heart valve lesion, with a prevalence of 1% to 2% among the general population, leading to severe regurgitation in ≈20% and aortic root/ascending aortic or aortic arch dilation in ≈60%. BAV is not uniform with respect to patterns of cusp fusion (or nonseparation). The Sievers and Schmidtke classification system makes a distinction among 3 main categories, depending on the number of raphes: S0 is a naturally perfect BAV without a raphe; S1 is the major type with a single raphe; and S2 has 2 raphes. There are 6 subtypes according to the location of the raphe(s) with respect to the commissure and 4 secondary subcategories according to the functional status of the valve: No for Normal, I for insufficient/regurgitant, S for stenotic, and B for both/mixed lesions. This classification system has proven to be useful for efficient and meaningful communication of cusp pathological anatomy.

The mechanism of AR in most BAVs, however, is a combination of dilation and distortion of aortic root geometry and of the functional aortic annulus, including a severely elliptical aortic annulus in some of the Sievers naturally perfect BAVs, with cusp pathology a combination of AR types I, II, and III. An additional issue that complicates surgical decision making in this subgroup of patients is the varying extent of distal aortopathy involving the aortic root, tubular ascending aorta, and aortic arch. Fazel et al have identified 4 main cluster categories of aortopathy in BAV disease to help approach this highly variable disease. Clusters I and IV involve the aortic root only (cluster I) and the aortic root and complete aortic arch (cluster IV). In aortopathy clusters II and III, the aortic annulus and the root are of normal diameter, and dilation starts at the supravalvular level, extending into the tubular ascending aorta (cluster II) or beyond the pericardial reflection into portions of the aortic arch (cluster III). It should be noted that clusters I and II are often combined (I+II, Ia, and Ib) because dilation of the aortic root, dilation of the sinotubular junction, and dilation of the tubular ascending aorta are often found in combination.

In the major type of pathological cusp configuration in BAV, a fusion of the left and right coronary cusps with 1 raphe, the noncoronary sinus is often enlarged. In addition, the insertion...
line of 1 of the cusps, often the noncoronary, is relatively long, which leads to prolapse of the noncoronary cusp into the left ventricular outflow tract (AR type II). The fused left and right coronary cusps also are often prolapsed as a result of cusp tissue redundancy. An overly long cusp insertion line can also lower cusp mobility (AR type III). Another mechanism of AR in BAV is a restrictive calcified or fibroed raphe, resulting in reduced cusp mobility or a free-margin tissue defect at the site of the raphe.

In BAV disease with a salvageable valve, treatment of AR in most cases should include replacement of the aorta and remodeling or reimplantation of aortic sinus tissue, in combination with correction of cusp pathology. Repair strategies that are used in BAV are free-margin plication, triangular raphe resection or shaving of the raphe, and free-margin resuspension. Another less frequently reported technique includes creation of a pericardial neocusp, as described by Urbanski.

**Risk Factors: Why Do Repaired Valves Fail?**

Different independent risk factors for failure of aortic valve repair have been identified.0,21 The presence of persistent AR immediately after valve repair, as assessed by intraoperative or early postoperative transesophageal echocardiography, is a strong predictor of subsequent progression of AR. Rates of recurrent AR and repeat surgical intervention also depend on the coaptation height of the repaired valve cusps: the height of apposition of the free margins and lunulae (the upper third of the coaptation height of the repaired valve cusps: the height of recurrent AR and repeat surgical intervention also depend on the coaptation height of the repaired valve cusps: the height of apposition of the free margins and lunulae). If the coaptation height is <4 mm, there is a higher likelihood of failure of aortic valve repair. The goal should be a coaptation height between 8 and 10 mm, as immediately validated by intraoperative transesophageal echocardiography. A coaptation level below the level of the aortic annulus is another powerful predictor of aortic valve repair failure (odds ratio, 7.9 for recurrent AR), equivalent to a postrepair mitral valve coaptation level above the mitral annulus.

Furthermore, Nash et al2 have identified preoperative factors favorable for the long-term success of valve repair: an eccentric jet of AR, absence of cusp or commissural thickening, and lack of cusp calcification. The last 2 factors intuitively seem to be related to the absence of the need for extensive cusp reconstruction, allowing a repair involving only plication of thin, fine, and pliable cusp tissue. The favorable impact of an eccentric AR jet is more difficult to explain because an eccentric jet is more likely in valves with a small range of free commissural aperture orientation and has been linked to higher rates of structural valve deterioration and reoperation.23

Finally, the number and location of raphes and the extent of their fusion may affect the late success of reconstructive operations for BAV. BAVs with larger angles and complete cusp fusion have been linked to favorable functional outcomes. In their 2011 series from Homburg, Germany, Aicher et al23 found that the absence of a root replacement procedure and the use of subcommissural plication and pericardial patch implantation had a negative impact on the durability of valve repair. Subannular plication has no demonstrable positive effect on outcome in patients with a large aortoventricular diameter, suggesting that root replacement in addition to cusp repair is likely to be a better option for these patients.

**Results**

The results reviewed and discussed here have been reported in publications of several larger clinical, prospective, and retrospective analyses. For this article, criteria for study selection were completeness of reported results, including structural valve deterioration and reoperation, formal report of conclusive survival and morbidity/mortality rate, substantial case numbers, and experience of the respective surgical center.

A series published by Aicher et al16 in 2004 reported 60 patients with aortic root remodeling in the setting of a BAV and an additional 130 patients who underwent root remodeling in the setting of a TAV. Repair of cusp prolapse was performed substantially more often with BAV (50 of 60 patients) than with TAV (47 of 130 patients). In addition, Alsoufi et al24 reported their experience with 71 patients followed up for a mean of 3.5 years after BAV repair, either isolated or in conjunction with aortic root remodeling or reimplantation procedures (41% of cases). In addition to free-margin plication, triangular raphe resection, and subannular plication, free-margin reinforcement cusp repair was undertaken in 14% of cases (with a double layer of Gore-Tex), as well as remodeling of single and multiple sinuses. In 2008, Pettersson et al from Cleveland, OH, reported 63 BAV and unicusp aortic valve patients, of whom 42 underwent an aortic valve repair procedure. Concomitant to their introduction of a repair-oriented classification of AR, Boodoo et al25 reported 264 patients in 2009 (90 BAV and 171 TAV) who underwent elective aortic valve repair with a median follow-up of close to 4 years, with details on mechanisms of AR: 80 patients had 2 contributing lesions, and 16 had 3 different mechanisms contributing to severe AR. In their update series on BAV alone, Boodoo et al26 presented results from 122 patients in 2010 who were recruited between 1995 and 2008.

Aicher et al25 summed up a total of 12 years of experience in a 2010 report on 640 patients after aortic valve repair for severe AR: 205 had BAV and 411 had TAV. Cusp repair was performed in 529, and 208 underwent a combination of cusp repair and a root procedure. The duration of follow-up was extended to 10 years.

Ashikhmina et al27 reported in 2010 a retrospective analysis of 108 BAV patients undergoing aortic valve repair (56% isolated), with a mean follow-up of 5.1 years. A matched-pair analysis was undertaken (81 matches) to identify differences in outcome between aortic valve repair patients and those who underwent bioprosthetic valve replacement. Badio et al28 from Munich, Germany, reported in 2011 a composite TAV and BAV population of 100 patients (80 male and 20 female patient; 43 BAV) aimed at showing differences in midterm outcome between those 2 subcohorts for aortic valve repair. The mean follow-up was 22 months with a cumulative follow-up of 167 patient-years.

The Table shows results of the selected studies on BAV and TAV repair.
Table. Outcomes as Reported in Recent Series on Bicuspid and Tricuspid Aortic Valve Repair: Freedom From Various Adverse Outcomes

<table>
<thead>
<tr>
<th>Author</th>
<th>Location, Country</th>
<th>Year</th>
<th>Root Management</th>
<th>Bicuspid AV Repair</th>
<th>Tricuspid AV Repair</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Patients, n</td>
<td>Reoperation at 5 y, %</td>
</tr>
<tr>
<td>Aicher et al16</td>
<td>Homburg, Germany</td>
<td>2004</td>
<td>Remodeling</td>
<td>60</td>
<td>2</td>
</tr>
<tr>
<td>Alsoufi et al24</td>
<td>Toronto, ON, Canada</td>
<td>2005</td>
<td>Remodeling, reimplantation</td>
<td>71</td>
<td>10.1</td>
</tr>
<tr>
<td>Pettersson et al4</td>
<td>Cleveland, OH</td>
<td>2008</td>
<td>Supracoronal ascending replacement, wedge ascending aortoplasty</td>
<td>41</td>
<td>9.5</td>
</tr>
<tr>
<td>Boodhwani et al28</td>
<td>Brussels, Belgium</td>
<td>2009</td>
<td>Annuloplasty of the AVJ and STJ with or without prosthetic aortic replacement</td>
<td>90</td>
<td>8*</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Aicher et al25</td>
<td>Homburg, Germany</td>
<td>2010</td>
<td>Isolated AV repair, remodeling</td>
<td>205</td>
<td>12</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Ashikhmina et al26</td>
<td>Rochester, MN</td>
<td>2010</td>
<td>Ascending graft replacement, reduction, aortoplasty</td>
<td>108</td>
<td>11</td>
</tr>
<tr>
<td>Boodhwani et al2</td>
<td>Brussels, Belgium</td>
<td>2010</td>
<td>Reimplantation</td>
<td>122</td>
<td>6</td>
</tr>
<tr>
<td>Badiu et al7</td>
<td>Munich, Germany</td>
<td>2011</td>
<td>Remodeling, reimplantation</td>
<td>43</td>
<td>15</td>
</tr>
<tr>
<td>De Kerchove et al10</td>
<td>Brussels, Belgium</td>
<td>2011</td>
<td>Reimplantation, subcommissural annuloplasty</td>
<td>161</td>
<td>0/10</td>
</tr>
<tr>
<td>Svensson et al6</td>
<td>Cleveland Clinic Foundation, Cleveland, OH</td>
<td>2011</td>
<td>Modified David, remodeling, STJ tailoring</td>
<td>366</td>
<td>8</td>
</tr>
</tbody>
</table>

Reoperation includes redo cusp repair and reoperation for prosthetic AV replacement. Three-year and 8-year data are listed if reported in the original articles instead of 5 and 10 years of follow-up, as indicated. AV indicates aortic valve; AVJ, aortoventricular junction; NA, not applicable; and STJ, sinotubular junction.

*Information not stratified for bicuspid and tricuspid AV; total numbers for composite patient population are given.
Survival

Better survival (Figure 3, comparative data) of the patients with BAV as opposed to patients with TAV anatomy was found in several studies. Aicher et al report a survival of 100% after 1, 3, and 5 years for the BAV patients, whereas survival was 93%, 87%, and 82% for the TAV group. However, there were important differences in clinical variables between the groups: a substantial difference in age and less frequent need of emergent procedures and arch replacement in the BAV group and lower rates of acute dissection but more frequent need of concomitant coronary artery bypass graft procedures. Overall 30-day mortality was 4.2% in the Homburg series; all had TAV, and almost half had acute aortic dissection. No hospital deaths were directly related to function of the repaired valve. Reported causes of death were mesenteric ischemia, sepsis, cardiac failure, and pulmonary embolism. In the Aicher et al updated series in 2010, only 1 patient died in the hospital after isolated aortic valve repair.

Overall survival in the 264 patients in the aortic valve repair series reported by Boodhwani et al was 95% at 5 years and 87% at 8 years. Alsoufi et al reported even lower mortality rates, with only 1 death (resulting from a noncardiac cause) 8 years after BAV repair. In the Mayo Clinic series reported by Ashikhmina et al in 2010, no significant difference in survival was found among BAV patients after bioprosthetic valve replacement versus aortic valve repair. After valve repair, the survival curves were similar to those of an age- and sex-matched population (actuarial 1-, 5- and 10-year survival rates were 99%, 96%, and 87%, respectively). In their updated series on BAV repair, Boodhwani et al reported no operative mortality. Overall mortality rate at 10 years was 3%, substantially different from the 13% in the earlier series in a combined BAV and TAV patient population. This again suggests better overall survival of the BAV than the TAV cohort after aortic valve repair.

No operative deaths were reported by Badiu et al, but 3 TAV patients died postoperatively as a result of cardiac failure;
underwent an emergent operation, and 1 succumbed to pulmonary complications. In this series, there was no difference in survival for the TAV and BAV groups, and overall estimated 3-year survival was 93%.

**Valve Function and Reoperation**

At the time of early postoperative evaluation, pressure gradients and other hemodynamic characteristics of the reconstructed BAV seem to be comparable to those of normal tricuspid controls.  

The severity of AR was markedly decreased after repair, from 2.5 to 0.8 postoperatively for BAV and from 2.8 to 0.9 postoperatively for TAV patients. Aicher et al. however, report subsequent progression of AR in a substantial proportion of patients: 6% of BAV and 8.5% of TAV. Four patients (2%) required reoperation for severe recurrent AR; 3 were in the TAV group. Actuarial freedom from recurrent AR at 1, 3, and 5 years was 98%, 96%, and 96% in the BAV patients and 93%, 89%, and 83% in the TAV patients. Twelve percent of BAV patients had undergone reoperation by 5 years and 19% by 10 years; in those with TAV, 3% and 7% had undergone reoperation for recurrent AR at 5 and 10 years.

In the Pettersson et al. series, the reasons for a relatively high rate of reoperation (4 of 42 patients who had undergone aortic valve repair) were dehiscence of the commissural realignment, torn cusp plication sutures, and rupture of a suture for fused cusp repair. Three patients were in need of subsequent valve replacement, whereas in 1 patient, a repair could be performed. In addition, 3 patients (4.8%) developed >3+ AR postoperatively while still in the hospital and had to be taken back to the operating room for valve replacement.

AR recurrence (Figure 4, comparative data) in the series reported by Boodhwani et al. was considerably higher for patients with type III AR (hazard ratio, 2.6; 95% confidence interval, 1.1–11.6), suggesting that cusp retraction and lowered mobility add to the risk of recurrent AR. The presence of AR grade >2+ was 12% at 5 years and as high as 21% at 8 years of follow-up. In their updated 2010 series, Aicher et al. were able to show a significant impact of experience in improving valve competence over time by stratifying the repair groups into sets depending on the year of operation. Similarly, Ashikhmina et al. were able to show a lower risk of reoperation (replacement) in patients who had valve repair after 2000. Although the difference in reoperation rates between the BAV and TAV population—higher for BAV—was significant, the replacement rate was similar: 13 of 36 bicuspid valves could be repaired.

Boodhwani et al. report freedom from recurrent regurgitation of 94% at 5 years, which is a remarkably good result in a pure BAV patient population and contrasts with the results from Homburg, Germany. Subgroup analysis in the Boodhwani et al. BAV series revealed greater freedom from recurrent AR for patients who had root replacement, either remodeling or reimplantation, in contrast to patients who had subannular plication or supracoronary aortic graft replacement. This finding underlines the importance of El Khoury’s emphasis on an exact definition of structural causes of AR and the importance of addressing each lesion contributing to AR: cusp pathology and geometric changes of the aortic root and functional aortic annulus (aortoventricular junction and sino-tubular junction).

It has recently been proposed that valve configuration, including completeness of cusp fusion and circumferential orientation of the free (nonfused) commissures, has an impact on late valve function and on the subsequent need for reoperation. Aicher et al. reported a difference in outcome for the different circumferential free commissural orientations in BAV. Individuals were grouped into a >160° and <160° group. In univariate analysis, patients with a free commissural orientation <160° required reoperation significantly more often (hazard ratio, 0.96; P=0.002); incomplete fusion of cusps was also linked to worse functional outcome.

No difference in reoperation rates (Figure 5, comparative data) between the TAV and BAV groups was found by Badiu and colleagues in their 2011 series. Estimated overall 3-year freedom from reoperation was 86%. In this series as in others, patients who underwent the reimplantation procedure to treat root dilation fared significantly better in terms of freedom from reoperation: 14% of TAV patients without any root procedure required reoperation within 3

**Figure 5.** Parametric survival estimation (freedom from reoperation) for patients with bicuspid and tricuspid aortic valves and Marfan syndrome after root reconstruction with or without repair. Data taken from Reference 27. AR indicates aortic regurgitation.
years, whereas none of the TAV patients with additional David reimplantation procedures needed reoperation during the follow-up period.

Other Adverse Outcomes: Thromboembolism and Endocarditis

The rates of other early-term and midterm adverse outcomes like thromboembolism and endocarditis were relatively low overall. In the 2004 Aicher et al\(^{16}\) series, neither complication was noted; the 2010 update\(^{25}\) showed low rates for both endocarditis (0.16% per patient-year) and thromboembolism (0.2% per patient-year).\(^{16,25}\) Alsoufi et al\(^{24}\) report higher numbers: 90% freedom from aortic valve endocarditis after 8 years, with 2 patients requiring reoperation for aortic valve repair. In the Toronto series of 2005, only 1 perioperative stroke was reported, and freedom from late thromboembolism or hemorrhage was 100% at 8 years of clinical follow-up. Pettersson et al\(^{24}\) reported no thromboembolic or infectious events with a median follow-up of 13 months. Boodhwani et al\(^{22}\) reported 4 strokes during follow-up (1.5%), 1 transient neurological event, and 1 instance of aortic valve endocarditis (each 0.3%); in the BA V update series, a linearized rate of 0.6% per patient-year for thromboembolic events was calculated.\(^{2}\) There was no difference between the TAV and BA V groups; 3-year freedom from neurological events or other types of thromboembolism was 94% for TAV and 97.9% for BA V.

Conventional Treatment: Valve Replacement

Many of the bicuspid types of aortic valves, especially in the setting of an additional stenotic component most often as a result of reduced fused cusp mobility, are often treated by prosthetic valve replacement. The same is still true for many severely leaking (aortic insufficiency, +4) BA V s or TAV s or very complex mechanisms of AR. These conventional procedures include biological or mechanical aortic valve replacement and composite valved graft procedures with reinsertion of the coronary arteries. Ten years after biological valve replacement, 25% of patients <65 years of age experience structural valve deterioration, and about the same rates of reoperation were observed.\(^{29}\) Survival at 10 years is \(\approx60\%\) without any difference between biological and mechanical prostheses. Event-free survival, including reoperation, stroke, or bleeding, after composite valved graft procedures has been reported to be as high as 78% at 8 years.\(^{30}\)

Discussion

Aortic valve repair is currently considered an attractive option to treat the regurgitant BA V or TAV. This seems to be true especially for the young patient with BA V, who often presents with qualitatively normal valve cusps, prolapse resulting from cusp tissue redundancy, and root dilation or a severely elliptical or dilated annulus. Low rates of systemic and central nervous system thromboembolism, infectious endocarditis, and hemorrhagic events are unquestionable benefits of a reconstructive approach to the regurgitant aortic valve.

However, BA V repair is accompanied by a considerable rate of recurrent and progressive AR. The need for reoperation has been reported to be as high as 51% after 10 years,\(^{26}\) in the 2010 Homburg BA V series,\(^{16}\) every fifth patient required reoperation after 10 years. A highly selective policy for including cusp repair may be advisable, acknowledging the fact that restricted cusp motion, cusp tissue defects, or calcification, among other cusp abnormalities, leads to higher rates of failure with valve-sparing surgery. Complex cusp lesions should be repaired only by surgeons with expertise, given the evidence that experience has a significant impact on the midterm and long-term success of valve repair. Figure 6 shows a decisional diagram for patient selection.

Aortic valve repair and V-SARR in the setting of an acute complicated type A dissection is still controversial.\(^{31}\) New devices and mechanical reconstruction adjuncts are now being introduced to the market, and their usefulness and results will have to be closely followed up over the next few years.\(^{32}\) A new sinus prosthesis that can probably maintain physiological root anatomy is available;\(^{33}\) widespread use can be expected once conclusive midterm and long-term results on functional valve outcome are available. In addition, an aortic annuloplastic ring for annular downsizing and stabilization\(^{34,35}\) may prove helpful in numerous reconstructive approaches to treat the leaking aortic valve, and its use also may increase in the future. In vitro evaluation of this device has delivered promising initial results.\(^{36}\)

To ensure durable repair of a severely leaking aortic valve, aortic root management must be chosen carefully. There are data suggesting that reimplantation may result in the most long-lasting repairs and that subannular reconstruction is less satisfactory. The reported results suggest that both downsizing the annulus and providing annular plication are crucial for satisfactory midterm repair in a substantial number of patients, especially those with large annuli or Marfan syndrome.

Reoperation rates tend to be higher for repair of BA V than TAV. There are some data suggesting that valve replacement rates do not differ because a considerable proportion of bicuspid valves can be rerepaired, but there is little evidence for the early and late surgical results of repeat valve repairs. To answer the question of whether the benefits of an initial reconstructive approach outweigh the risks accompanying subsequent reoperations, additional comparative long-term studies are required.

Reconstructive valve surgery translates into convincing short- and medium-term survival rates. Long-term results are sparse and not yet conclusive. Although a significant difference has not been reported, BA V patients tend to have a better medium-term survival than TAV patients after reconstructive aortic valve surgery, most likely because of substantial differences in clinical variables and patient characteristics. A large proportion of the postoperative mortality seems not to be related to worsening function of the valve or to other cardiac causes. Survival rates after aortic valve repair in both groups, however, have been shown not to be substantially different from survival of age- and sex-matched general populations. However, no improvement in survival after valve repair as opposed to bioprosthetic valve replacement can be demonstrated.

The option of catheter-based valve-in-valve placement for late valve stenosis after bioprosthetic aortic valve replacement seems to favor bioprosthetic replacement. The stenotic lesion resulting from a bioprosthetic root or valve calcific degeneration seems to be a much more convenient setting for proper
placement of a transcatheter prosthesis as opposed to the regurgitant valve after V-SARR with pathologically normal cusps.

Different aspects make it difficult to compare the treatment modalities of prosthetic aortic valve replacement with repair of the native aortic valve and to draw solid conclusions. The adverse events to be expected after aortic valve repair—worsening AR, structural valve deterioration requiring reoperation, thromboembolism, and endocarditis—are generally rare and late events. A prospective randomized study would be prohibitively expensive and therefore is unlikely to be performed. Thus, evidence needs to be generated by meta-analyses of reported observational studies. Long-term results for TAV and BAV repair are not yet available; therefore, at present, patient selection should remain restrictive and reconstructive procedures should be performed only by highly experienced teams despite encouraging short- and medium-term outcomes.

Conclusions
A high degree of freedom from thromboembolism, bleeding, and infectious complications favors valve reconstruction over replacement in a substantial cohort of selected patients. The overall survival after TAV and BAV repair is high, with better survival for the BAV cohort, albeit confounded by clinical variables. Substantial AR progression has been shown in some reports, and data indicate that reimplantation in conjunction with valve repair is more durable than isolated cusp repair or cusp repair with aortic root remodeling. To achieve a durable result, it is necessary to analyze every mechanical contributor to AR preoperatively and to approach every lesion in a combined replacement (sinus tissue) and reconstruction attempt (other components of the aortic root: cusps, commissures, aortoventricular junction, sinotubular junction). In BAV repair, there is an increasing understanding of the impact of valve configuration on long-term durability, including the completeness of cusp fusion, free commissural orientation, and presence or absence of a raphe (Sievers type of BAV configuration). Valve function late postoperatively is linked to the year of the procedure and surgeon experience, indicating a long, steep learning curve for this technically unforgiving procedure.

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None.

References


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Fabian A. Kari, Matthias Siepe, Hans-Hinrich Sievers and Friedhelm Beyersdorf

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